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ABSTRACT

The authors describe personal experiences with Usher's Syndrome, a condition involving visual impairment (due to Retinitis Pigmentosa) and deafness. Prevalence and onset of Usher's Syndrome are described along with characteristics of two different types. Problems connected with diagnosis are reviewed, including genetic aspects and the progressive nature of the visual loss. Types of ophthalmological techniques are described, including oculomotor tests, retinoscopy, and electroretinography. Five stages of the syndrome are noted: (1) awareness, (2) general counseling, (3) general planning and community resource identification, (4) specific planning and adjustment counseling, and (5) adjustment. Service implications are reviewed along with 31 common behaviors characteristic of Usher's Syndrome children. (CL)

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USHER'S SYNDROME:

A 1986 Update

by

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and

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May, 1986

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I grew up on a farm in Tennessee, a deaf child of hearing parents. In those days nobody outside of specialized medical circles had ever heard of Usher's Syndrome. During my early years at the Tennessee School for the Deaf in Knoxville, I was not aware that something may have been wrong with my vision. At about age ten, I became unconsciously aware that something was amiss. For instance, I began to notice that my schoolmates complained that it was often hard for them to attract my attention when they waved their arms. They kept suggesting that I ought to wear glasses. I also began to note that bumping into things and people, to which I had become more or less accustomed, was not something everyone did frequently. I found myself being blamed for little incidents that I unwittingly got involved in. All those things and bruises began to add up. It became obvious that something was not right, but I did not have the slightest idea how to define it. At about age 15, a deaf teacher helped me put a label on it. She called it tunnel vision. From then on, my schoolmates were more understanding about my problem and no longer got mad if I bumped into them, or if they failed to get my immediate attention. As I grew older, I became more experienced in avoiding those little embarrassing incidents.

I went through a succession of eye examinations from the time I was 10 until I was 25 to find out what was wrong and what might be done medically, but all the doctors said that there was nothing to worry about, so I believed them. At age 36, I was finally diagnosed (Larson, 1985).

Usher's Syndrome. Chances are many of you did not relate

that term to the story I just read. Today, my goal is to enlighten and inform you of this very prevalent syndrome, which I believe all of you should become very aware of. Not only for your sake, but for our children's.

According to Vernon, et al. (1982), the combination of deafness and retinitis pigmentosa was first reported by von Graefe in 1858, but Usher, a British ophthalmologist, was first to stress the hereditary nature of the disorder which now bears his name (Usher, 1914). Unfortunately it took 115 years to progress from the discovery of the disease to the first coordinated multidisciplinary effort to prevent its occurrence and provide rehabilitation for those who have it. It is almost unbelievable that a condition which causes 3 percent to 6 percent of deaf children to become blind is still relatively ignored by professionals working with deaf adults and children (Vernon, 1974).

Presently there are 16,000 people with this disease in the United States. Out of the general population 3/100,000 people are inflicted with Usher's Syndrome.

According to Judy M. Pierce (1986), there are presently two types of Usher's Syndrome today. The following characteristics represent the different types.

Type One

--profound hearing loss, 100 dB or # greater;

--no intelligible speech;

--vestibular system disturbed;

--night blindness appears around the age of twenty

--profound field loss

--flat ERG.

Type Two

--hearing loss of moderate to profound

--person exhibits nystagmus

--night blindness appears around age thirty or forty

--visual field varies from normal to marked

--in ERG-some electricity

Usher's Syndrome is a genetic condition involving hearing loss and gradual progressive blindness due to retinitis pigmentosa. Retinitis pigmentosa (RP) is the name applied to a group of hereditary diseases causing degeneration of the retina. In Usher's Syndrome, the affected individual has both RP and a profound congenital hearing loss. While the loss of hearing is apparent near birth or in very early childhood, the vision impairments frequently remain undetected until late adolescence or the early twenties. The affected individual may notice some difficulties, but may not be consciously aware of other symptoms until an ophthalmologist asks quite specific questions about them (The National Academy, Gallaudet College).

Two major professional problems come to light when diagnosis is made. First, few of those who diagnose the retinitis connect it with the deafness. Consequently, the condition is not identified as Usher's Syndrome. Second, in addition to overlooking the presence of Usher's Syndrome, the person "making the diagnosis" is often unaware of the genetics of the disease or even of the fact that it is a recessive inherited

condition (Vernon, 1974).

In most instances it is inherited. The most common inheritance of RP is called "recessive." This means that two donors of the gene are needed to acquire RP, although the individuals involved may not have RP. Such individuals are known as carriers. When two carriers marry, the chance that a given child will be affected is one out of four.

Another form of RP inheritance is "sex-linked." In sex-linked RP the mother is the carrier. If a female carrier of sex-linked RP marries a normal male, there is a 50 percent probability her sons will be affected and a 50 percent chance her daughters will be carriers. Sex-linked RP is the one form in which carriers can often be detected.

The third inheritance pattern is called "dominant." In this type, affected persons have an affected parent, but unaffected children will have unaffected offspring. The recurrence risk for the offspring of an affected person is one in two, and males and females are affected in equal proportions.

Because RP runs in families, all family members are encouraged to have a thorough eye examination (RP Foundation, 1983).

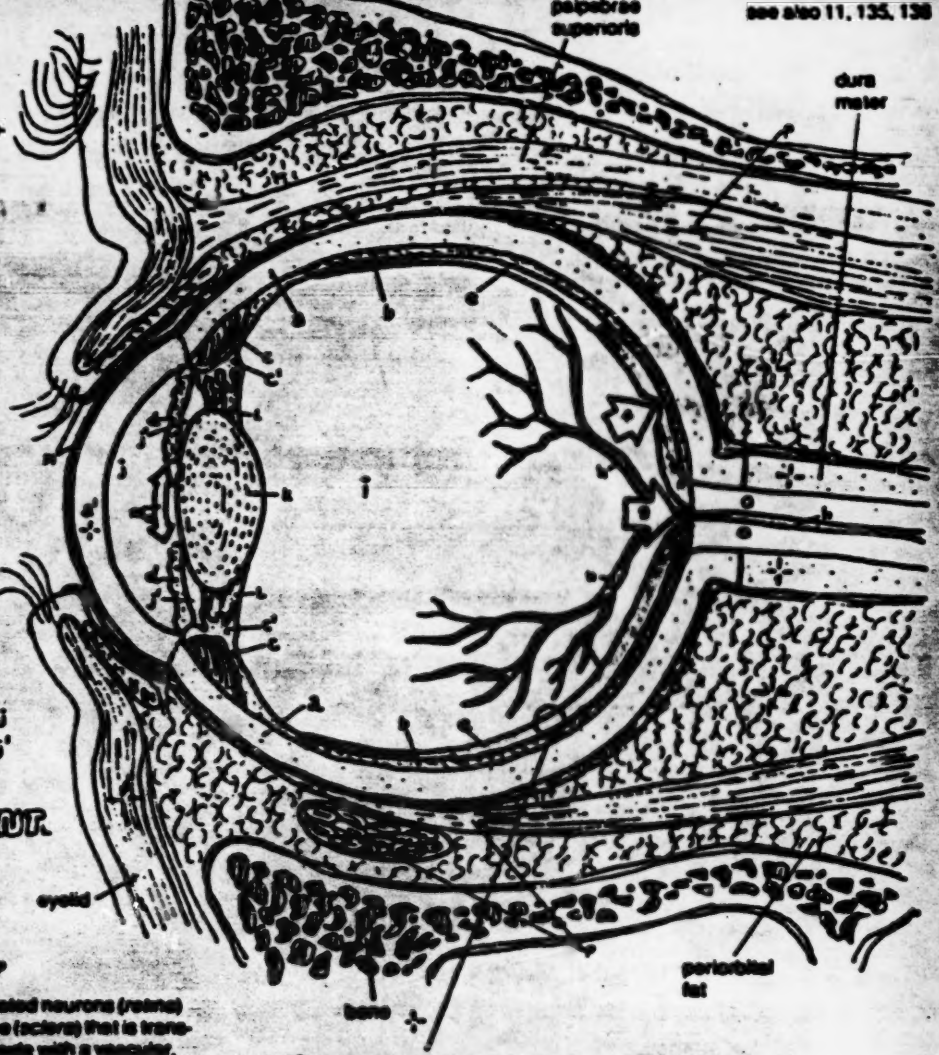
The symptoms of Usher's Syndrome are quite multiple in number. The degeneration of the retina by retinitis pigmentosa affects all or nearly all of the rods and cores of the retina. The rods and cores play important complimentary roles in the process of seeing: the rods are cells which enable the eye to perceive objects in dim light and darkness and also allow

VISUAL SYSTEM (1)

see also 11, 135, 138

ON 21

1. Name the nature of the neighboring structures of this vertical section of the eye which are not to be colored. The 3-dimensional character of the eyeball can be appreciated by looking at Plate 139.
2. Use a light color for i, j, and j'. Use red for h.



LAYERS OF THE EYE.
SCLERA.
CORNEA.
CHOROID.
CILIARY BODY.
CILIARY PROCESSES.
IRIS.
RETINA.
FOVEA CENTRALIS.
OPTIC DISC.
RETINAL ARTERIES.

SPACES / FLUIDS.
VITREOUS BODY;
AQUEOUS HUMOR;
ANTERIOR CHAMBER;
POSTERIOR CHAMBER;

LENS.
SUSPENSORY LIGAMENT.
PUPIL.
CONJUNCTIVA.
OPTIC NERVE.
MUSCLES OF EYEBALL.

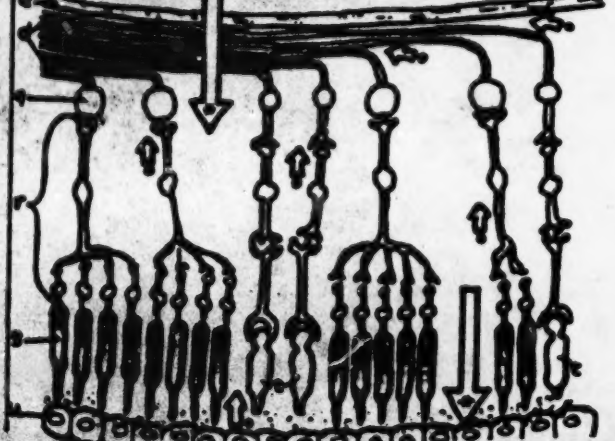
The eye is a layer of photoreceptor cells and associated neurons (retina) packaged within a fibrous, rubberlike protective globe (sclera) that is transparent in front (cornea). The sclera is lined on the inside with a vascular, highly pigmented layer (choroid) that absorbs light and prevents scattering. Most of the other structures within the eyeball are concerned with light waves. Light, traveling in straight lines, must be bent (refracted) as it enters the eye (through the pupil) to converge on a focal point in front of the retina. The cornea (a layer of fine fibers encapsulated with epithelial cells), and highly pigmented epithelial cells with a high degree of elasticity), vitreous humor (a gel of water and mucoprotein), and aqueous humor (an extracellular fluid filling the anterior/posterior chambers of the eye) are the media that refract the light waves, listed here in order of their functional significance.

The choroid thickens anteriorly to form the ciliary body (pigmented connective tissue with smooth muscle fibers) from whose processes the suspensory ligaments are directed to the lens. These ligaments passively put a tension (circumferentially) on the lens, flattening it. The ciliary muscle of the ciliary body, acting under parasympathetic stimulation, relaxes the tension of the ciliary processes and suspensory ligaments, resulting in the lens rounding up due to its inherent elasticity and permitting close-up vision.

The iris is an extension of the ciliary body and forms a circular, adjustable rim about the lens. The space within the circular rim is the pupil. The diameter of the pupil may be altered by muscles in the iris that are arranged in two directions. The dilator pupillae muscle pulls the iris back toward the ciliary body, dilating the pupil; the sphincter pupillae muscle drapes the iris down around the front of the lens, constricting the pupil. The color of the iris is largely a function of the amount of pigment within.

The retina consists of several layers of cell bodies (nuclear layer) and fibers (plexiform layers). The rods (highly sensitive to light and insensitive to color) and cones (sensitive to color and provide highest visual acuity) constitute the photoreceptor neurons; the rods generally have a population edge of 20:1. An ophthalmoscope here assure at the fovea centralis (a slight depression in a yellow, circular area of the retina), where only cone cells and related neurons appear. This point on the retina is in the optical pathway of direct light; thus the greatest visual acuity can be gained by looking directly at an object, under proper light. The optic disc (where optic nerve fibers stream to the outside behind the eyeball) is without rods/cones and is, in fact, a blind spot.

LAYERS OF RETINA.
INTERNAL LIMITING MEMBRANE.
AXONS (OPTIC N. FIBERS).
GANGLION NEURONS.
BIPOLAR NEURONS.
PHOTORECEPTOR NEURONS.
RODS. CONES.
PIGMENTED EPITHELIUM.
LIGHT RAYS. NERVE IMPULSE.



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for peripheral (side) vision; the cores enable the eye to see in the light and perceive color. The cores also control central (forward) vision. In the course of the disease, the functions of the rods and cores are impaired. There are three significant symptoms which are common to retinitis pigmentosa: 1) poor dark adaptation, 2) poor side vision and 3) poor central vision.

During poor dark adaptation, the individual has difficulty seeing at night or in dark places. Slow or incomplete adjustment to the dark is usually present during childhood, but is not "officially" detected until later. In some cases, poor dark adaptation is referred to as "night blindness."

When experiencing poor side vision, the individual cannot perceive objects in the normal peripheral region; this is also called "visual field loss" or "tunnel vision." It is a progressive loss, the vision becoming more truncated over time. Poor vision is usually, present by the late teens. However, because central vision is still relatively good and the individual can compensate for the field loss by moving his eyes, it may not be noticed until it is fairly advanced.

Poor central vision is when the distance of the individual's central vision is limited. This loss, too, is progressive; the individual sees fewer and fewer details in the distance over a period of time. Poor central vision is generally not present until the individual has reached the late twenties or thirties, and usually does not decline to the point of total blindness until much later in life. Some people retain a few feet of central vision all of their lives. But it should be noted that

with a very small visual field, even good central vision is of limited use (The National Academy).

In the identification of Usher's Syndrome, there are many technical and highly sophisticated ophthalmological techniques used today.

According to Wanda Hicks (1978), objective tests and procedures are by definition those evaluation methods which do not require a cognitive response by the patient, so patient subjectivity is eliminated. The objective tests to be discussed here will include: 1) oculomotor tests; 2) ophthalmoscopy; 3) retinoscopy; 4) tonometry; and 5) electroretinography.

Oculomotor Tests--These are designed to determine the presence or absence of normal pupillary reflexes (their response to light, etc.) as well as to quickly assess the range of eye movements. Such procedures are valuable, not only for the assessment of visual function, but also for the detection of some serious nurological conditions. They are quick and easy to do and should always be part of a vision screening. They do not, however, have any primary value in the diagnosis of Usher's Syndrome.

Ophthalmoscopy--This procedure is one which employs a hand held optical device called an ophthalmoscope to get a better look at the eye. While it is most often used to look at the back of the eye (the fundus), it can also be used to get a magnified view of other eye structures. In a screening, the emphasis in ophalmoscopy is placed on detection of abnormalities rather than diagnosis per se.

Retinoscopy--Here again a hand held optical device is employed to look at the eye. The retinoscope employs a "streak" of light focused in a certain way to estimate the refractive error of a patient. In this way the screener can get a rough idea about the type and magnitude of the optical correction which may be required. As with ophthalmoscopy, a certain amount of experience is required by the screener for this technique to yield meaningful results. The technique has no primary value for the diagnosis of Usher's Syndrome.

Tonometry--By this procedure the internal pressure of the eye is measured. Normal pressures range roughly from 10-25 mm of mercury. When they go higher one becomes concerned about a condition called glaucoma. To measure internal pressures a device called a tonometer is used. There are many types of tonometers but they all must depress or flatter the eye (very slightly) with a known force in order for a calculation of pressure to be made. When the results of tonometry are evaluated it is important to realize that higher than "normal" pressures do not always mean that the patient has glaucoma. Tonometry has no primary value in the detection of Usher's.

Electroretinography--The light sensitive neural structure in the back of the eye is called the retina. It is actually an extension of the brain. It is the job of the retina to convert an optically formed picture of the world we look at into nerve impulses which are then transmitted to other parts of the central nervous system. This neural activity produces a very small amount of electricity which can be recorded

at the front of the eye. Such a record is called an Electroretinogram (ERG). Simple stated the ERG is a graphical representation of the change in voltage as a function of time following a flash of light in the eye.

The methods and procedures employed are well within the capabilities of an adequately trained technician. The patient must wear a contact lens electrode to which a wire is attached. Another "reference" electrode is taped to the skin beside the eye. These wires are all intended only to pick up tiny electrical currents normally present in the eye. No external source of current is ever passed through the eye (Hicks, 1978).

Those technical diagnosing instruments that have been described above are effective in making the diagnosis of Usher's Syndrome at later stages.

The stages of Usher's Syndrome may lead to a logical sequencing of educational programming, the degree of counseling needed and the stages may prove to be very beneficial in the area of rehabilitation.

This disease has been divided into five stages as follows:

USHER'S SYNDROME

Outline of Characteristics for
Program Consideration

I. Stage 1 - "Awareness Stage"

A. Period: Middle Childhood

(C.A. 6-9) to Pre-Adolescence

(C.A. 10-12)

B. Symptoms:

1. Congenital deafness;
2. Loss of some right vision.

C. Programming Approach

1. Comprehensive diagnosis and disease identification;
2. Describe initial symptoms of the disease to the child;
3. Alert the child to precautions and make recommendations regarding compensatory measures the child might take in coping with night problems;
4. Provide genetic counseling to the family.

II. Stage 2 - "General Counseling Stage"

A. Period: Early Adolescence (C.A. 13-16) to late adolescence (C.A. 17-20)

B. Symptoms:

1. Loss of night vision;
2. Some loss of peripheral vision.

C. Programming Approach:

1. Additional genetic counseling (family and individual);
2. Career counseling;
3. Specific counseling regarding the nature of the disease, phases of the disease, and variations in the rate of disease progression;
4. Educational program adjusted as necessary to accommodate the early symptoms of the disease.

III. Stage 3 - "General Planning and Community Resource Identification Stage"

A. Period: Early adult years.

B. Symptoms:

1. Loss of night vision;
2. Loss of most peripheral vision;
3. Minimal loss of central vision.

C. Programming Approach: Establishment (or maintenance) of a close liaison relationship with appropriate community/state/federal resource agencies. Identification of an agency or individual should be made who will assume primary responsibility for "case management." The case manager should "track" the individual and provide/arrange the services needed from the various on-call agencies. The agencies might include:

1. Pre- and post-high school programs attended by the individual;
2. Center(s) for continuing education;
3. Medical profession, including personal physician, clinics, and hospitals;
4. State vocational rehabilitation agencies;
5. National and State organizations for the deaf;
6. National and State organizations for the blind.
8. Speech and hearing agencies (clinical and evaluative services);
9. Civic organizations (including social and recreational groups);

10. Religious organizations;

11. Private rehabilitation agency.

IV. Stage 4 - "Specific Planning and Adjustment Counseling Stage"

A. Period: Middle adult years (could occur sooner or later).

B. Symptoms:

1. Loss of peripheral vision;

2. Significant loss of central vision.

C. Programming Approach: Adjustment counseling/training might include the following areas (including continued use of all agency and individual resources):

1. Compensatory occupational training for maintenance of present job;

2. Occupational retraining for new job;

3. Introduction to occupation within a sheltered workshop situation;

4. Community/continuing/adult education programs for social, recreational, and skill training;

5. Core mobility training;

6. Braille training;

7. Training for maintenance of communication skills;

8. Training to alternate modes of communication;

9. Counseling for social, and particularly family, adjustment;

10. Social/psychological therapy.

V. Stage 5 - "Adjustment Stage"

A. Period: Late adult years (could be sooner).

B. Symptoms:

1. Loss of night vision;

2. Loss of peripheral vision;

3. Complete loss of central vision.

C. Programming Approach: (Including continued use of all agency and individual resources):

1. Occupational training/retraining for:

(a) Maintaining skills required in present job;

(b) Developing skills required for a new job;

(c) Work in a sheltered workshop;

(d) Maintenance of work-related responsibilities in the home setting.

2. Day-to-day therapy for:

(a) Maintaining/building an adequate communication modality;

(b) Improving Braille and core mobility skills;

(c) Maintaining social contact;

(d) Maintaining psychological well-being;

(e) Developing coping skills in all aspects of life.

3. Alternate placement considerations:

(a) Home environment

(b) Nursing home;

(c) Public/private institution;

(d) Hospitalization (if concomitant medical

problems exist);

(e) Other.

* Adapted from Jack English, (1978), A Five-State Study of Usher's.

Syndrome Incidence and Service Implications, pp. 364-371.

Caution must be strongly emphasized, however, in viewing the outline; that is, the age of the symptoms, and the speed of progression from one phase to another vary greatly from individual to individual. Thus, it should be emphasized that the stages outlined in the appendix, though having relevant programmatic value, should be considered fluid in both duration and age of onset (English, 1978).

During the fifth stage when the is loss of night, peripheral and complete loss of central vision, steps and decisions must be made concerning the new form and technique of communication to be used.

Other communication considerations are; the hand over hand or guided-wrist; the alphabet in palm of hand; the glove execution or Braille. These methadologies depend on degree of loss, comfortability and executability and personal preference.

The personal side of Usher's patients is very complex. After the diagnosis, many steps follow. Genetic counseling, family history, explanations of the possible future and family and individual counseling procedures begins.

According to Carolyn Torrie (1978), the constant need to redefine previous values, attitudes, and feelings and the

pressing need to re-examine these emotions brings to the adolescent internal shifting, sorting, and change. This questioning of what is felt has an eroding effect to any sense of security that the individual may have developed in the past. This erosion, so frequent in adolescence, requires the individual to call upon his strengths to learn to cope with these new feelings or new perspectives of feelings and value system.

The storm and stress is the outward symptom of the condition. The newly held experiences, changes in social, physical and emotional systems, and the redefinition of ways to relate to the environment are the underlying motif of adolescence. These components of adolescence, when observed systematically, may provide one part of a matrix for assessing the needs of adolescents with Usher's Syndrome.

Thus with all of these new, confusing and frustrating feelings arising, new behaviors will develop. Common behaviors of Usher's Syndrome students are listed below:

1) They may appear quiet in a large group;
2) They frequently exhibit shy-type behaviors, particularly in group situations;
3) They may edge themselves to one side when placed in a group;
4) They frequently bump into objects in the room such as tables and chairs;
5) They are bothered particularly by bright lights and complain that glaring lights hurt their eyes;
6) They frequently fail to understand or miss group

instruction that is given by residential advisors or classroom teachers;

7) They are frequently last in completing group activities;

8) They exhibit symptoms of anxiousness in new areas;

9) They are frequently last to enter a room and may bump into other students and objects;

10) They may appear unconcerned and may fail to fully participate in group activities;

11) They frequently must rely on a friend for information;

12) They have repetitive behavior;

13) They may frequently have accidents at meal time with objects placed at their sides;

14) They express a desire to enter a room that will be darkened very early;

15) They have a tendency to avoid conversations in darkened areas;

16) They may choose to stay at home alone rather than be faced with the embarrassment of not being able to see in dark or poorly lit places;

17) They may appear awkward when exiting from the inside to the outside of a building;

18) They may avoid participating in outdoor sports when the sun is very bright;

19) They may request sunglasses whenever they are asked to participate in activities in the bright sunlight;

20) Without sunglasses, they are nearly always seen shading their eyes or squinting when in the sunlight;

21) They avoid walking or running fast in unfamiliar areas, particularly when there is bright sunlight or when they are in darkened areas;

22) They frequently hesitate at the top or bottom of stairs;

23) When walking down the street, they frequently stumble over curbs or slight elevations/depressions in walking surface;

24) They may hesitate when first meeting someone, or appear to ignore them;

25) They have difficulty reading in dimly lit areas;

26) When walking along a road at night, they may appear to stagger or lose their balance after an oncoming car has passed;

27) They may avoid social activities when such activities take place in dimly lit areas;

28) They may appear to ignore you if you are standing at their side;

29) They prefer to sit at the side of a large group;

30) They prefer conversation at distances of 4 to 6 feet;

31) They may appear to be constantly visually scanning a group.

The above behaviors, though possibly appearing somewhat bizarre to the untrained observer, are logical when considering the student's reduced peripheral vision, inability to adapt to substantial changes in illumination, night blindness, and impaired central vision (Hicks and Hicks, 1981).

There are many implications, questions and concerns related to Usher's Syndrome as one can plainly see. What can be done?

According to the RP Foundation (1985), there is no known cure for Usher's Syndrome, intensive research is currently under way. Nevertheless, it is of utmost importance that an accurate diagnosis be made. This must include special electrodiagnostic tests of retinal function, visual field testing, hearing testing, and a thorough ophthalmological examination. Next, genetic counseling should be otained to define the risks of other family members or future offspring be affected. Optical and non-optical low vision aids, including an array of electronic devices, prisms and telescopes, are available when a decrease in vision is experienced. Sign language instruction and other communication skills can be learned and hearing aids fitted. Personal, education and vocational counseling from those familiar with the disease can usually be obtained from community resources. Adaptive training skills, job placement and income assistance are also available through the State Department of Vocational Rehabilitation or Division for the Visually handicapped.

In closing, Usher's Syndrome is a very serious and complex disease. I hope I have obtained my goal; of enlightening and informing you concerning this topic. We, as professionals, must not be idle. We must be cognizant of our environment and moreover our very special

children. Please be aware of your children's behavior, characteristics and course of development. Early intervention is the key to lessening this sad, but very real occurrence of letting this disease continue unnoticed.

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